Stormy onset with prolonged loss of consciousness in benign childhood epilepsy with occipital paroxysms

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Abstract
In nine of 62 children with benign occipital epilepsy (BOE) the onset was stormy and alarming. The first and often only seizure was characterised by prolonged loss of consciousness lasting up to 12 hours, suggesting an acute cerebral insult. In all but one case there was a tonic adversion either of eyes alone or of both head and eyes which was interpreted as conjugate deviation. The other accompanying ictal motor phenomena were either partial or generalised convulsions. In five patients the seizure was heralded by a headache, and in five cases was accompanied by vomiting. The seizure began with visual symptoms in only one patient. The seizure occurred while awake in seven and during sleep in two. The age at onset was from 3¼ to 10 years. Interictal EEGs showed occipital discharges typical of BOE, and the clinical course was benign. In four cases a few partial or complex partial seizures recurred during subsequent anticonvulsant therapy, but in five cases seizures never recurred. Anticonvulsants were discontinued in five patients who remained free from seizures for one to 11½ years after withdrawal of treatment. Sudden coma in a child associated with focal features such as tonic deviation of the head or eyes or both may represent a benign seizure disorder.

Benign occipital epilepsy has been defined by Gastaut[1][2] as another form of benign partial epilepsy in children. He described seizures which included visual symptoms often followed by motor or psychomotor manifestations and sometimes terminating with postictal migrainous symptoms such as headache, nausea, and vomiting. The typical interictal EEGs consisted of high voltage occipital spike-wave paroxysms attenuated by eye opening. The diagnostic criteria included a normal neurological state and no abnormal neuroradiological findings. The course is benign and seizures cease in most patients before adulthood. The clinical and EEG features of BOE have been reported by others.[3–9] Camfield *et al*[10] reported four adolescents with basilar migraine, infrequent cerebral seizures, and the same interictal EEG findings as in BOE with a benign clinical course. In 1980 Panayiotopoulos[11] described a similar case arguing in favour of the epileptic nature of the syndrome. Newton *et al*[4] in

describing the clinical features of 16 children with occipital spike-wave complexes suppressed by eye opening, stressed the association of this EEG pattern with a wide range of clinical disorders, including basilar artery migraine, BOE, and lesional epilepsy.

In a previous report[6] presented at the 17th International Epilepsy Congress in 1987 we summarised the seizure patterns in 62 children fulfilling the electroclinical criteria of BOE formulated by Gastaut,[1] finding visual symptoms in 18 adversive seizures (almost all contraversive) in 22, headache in 24, and vomiting in 15. Prolonged alarming loss of consciousness lasting up to 12 hours was the presenting symptom of the first seizure in nine patients who we describe here.

Patients and results
During the years 1970–85, 62 children were diagnosed as having BOE at the seizure clinic and the EEG Laboratory of the Beilinson medical center. The seizure patterns in these children have been briefly described elsewhere.[6] Data from seven boys and two girls who had their first seizure with prolonged loss of consciousness are presented here (table 1). The age at onset ranged from 3¼ to 10 years. In all cases there was no family history of epilepsy or migraine. One patient had suffered a simple febrile convulsion at the age of 2½ years and had a family history of febrile convulsions. Five patients suffered from headache before the seizure: in three it lasted less than 20 minutes, in two it was prolonged, lasting from 12 to 48 hours and fluctuating in severity, and in one patient it was accompanied by vomiting. Loss of consciousness, whether preceded or not by headache, was sudden and prolonged, lasting from four to 12 hours and accompanied in all but one by a tonic deviation either of eyes or both head and eyes followed by bilateral clonic jerks in two, generalised tonic clonic convulsions in two, left hemiclonic in one, right tonic clonic in one, and right brachiofacial in one. The patient (case 6, see table 2) whose initial seizure was non-convulsive later experienced complex partial seizures. Vomiting occurred in four patients during the adversive stage and in one in association with the headache preceding the seizure. In only one patient did the seizure begin with visual symptoms followed by loss of consciousness and convulsions.

EEG findings
In one patient (case 8) EEG was performed during the prolonged seizure, and it showed

continuous bilateral spike and wave complexes, more prominent on the left side. In seven patients EEG was obtained within 24 hours after the initial seizure and in one (case 1) after five days. In three of them occipital spikes were seen in this first record whereas in the five others whose first EEG showed occipital slow activity occipital spikes were recorded in subsequent EEGs obtained between two days to six months later. These waking interictal EEG recordings showed high voltage spike-waves or sharp and slow waves in the posterior regions, in runs or almost continuously, attenuated by eye opening. In five the discharges were unilateral, in four bilateral with predominance of one side. In two patients (cases 2 and 9, table 1) a few brief generalised spike-wave discharges were recorded in some of the later tracings in addition to the occipital focus, and in one case (case 5) sporadic centro-temporal spikes were seen in addition to the occipital focus three years after the initial EEG.

CT findings

CT scans performed during the initial seizure in seven patients were normal in five; in the other two (cases 1 and 2) narrowing of one or both lateral ventricles was demonstrated. The course was benign in all nine patients, despite the stormy onset. In five of eight patients who received anticonvulsant therapy after the first seizure there was no recurrence of seizures. Four of them (cases 3, 7, 8, 9, table 1) are no longer taking medication (for 3·5, 8·5, 5·0 and 11·5 years respectively). Among the three remaining patients one (case 5, table 2) had two brief adversive seizures with vomiting and loss of consciousness on waking after reduction of medication six and 24 months later. One of these patients (case 6, table 2) had two brief complex partial seizures after eight and 14 months. The course of the third patient (case 2) is also presented in table 2. In the one patient who did not receive any medication after the initial seizure (case 1, table 2) a brief second seizure occurred four months later.

His course is detailed in the case report given below. None of the nine patients had a recurrence of severe prolonged seizures comparable to the first one.

Illustrative case reports

Case 1 This 9 year old boy, some 30 minutes after he had fallen asleep was found one night in a "coma", gargling and vomiting, with his eyes and head turned to the right. There were fine clonic jerks of the hands and feet, slight cyanosis, and a loss of sphincter control. On transfer to our hospital he was unconscious with right hemiparesis and a right Babinksi sign. CT scan showed a narrow left ventricle due to oedema of the left hemisphere. He was intubated and given intravenous dexamethasone, mannitol, and diazepam. Several hours later his body temperature was 38·5°C, and acyclovir was started with the suspicion of encephalitis. The child regained consciousness after 10 hours with no neurological deficit. Five days later a repeat CT scan was normal, and the antiviral treatment was stopped; two EEG recordings made on the fifth and seventh days demonstrated a slow wave focus over the left occipital area. Four months later he experienced two brief nocturnal seizures, half an hour apart, with right tonic deviation of head and eyes, vomiting, and loss of consciousness. EEGs obtained at 12 hours and one week later showed repetitive spike and slow wave discharges over the left occipital region, inhibited by eye opening. Treatment was begun with 90 mg phenobarbitone daily. One and a half and two months later he had two additional adversive diurnal seizures without vomiting. In the former the loss of consciousness lasted 10 minutes with gradual return to full consciousness over 20 minutes, and in the latter it lasted for five minutes. Both seizures were associated with headache which preceded the seizure and continued post-ictally for eight and four hours respectively. EEG recordings made two, six, 12, and 16 months after the cessation of seizures showed the same abnormalities, but

Table 1 *Clinical data for nine children with benign childhood epilepsy with occipital paroxysms*

Case No	Age at onset (years)	Sex	Seizures when awake (W) or asleep (S)	Headache	Vomiting	Adversion	Other motor features	Duration of loss of consciousness (hours)	Interictal EEG	Duration of drug therapy (years)	Recurrence of seizures	Follow up (years)
1	9	M	S	No	Yes	Right head & eyes	Clonic jerks (all limbs)	10	Left occipital	Pb/3	Yes	4
2	3¼	M	W	Yes	No	Left eyes	Clonic jerks (all limbs)	12	Right > left occipital & generalised S/W	PHT/4	Yes	4
3	8	M	W	Yes	Yes	Left eyes	Left hemi-clonic	6	Right occipital	PHT/2	No	5½
4	4	M	S	No	Yes	Left head & eyes	Generalised tonic-clonic	4	Right > left occipital	CBZ/3½	No	3½
5	5	M	W	Yes	Yes	Left head & eyes	Generalised tonic-clonic	4	Right > left occipital	CBZ/3½	Yes	3½
6	8	M	W	Yes	No	None	None	5	Left occipital	CBZ/4	Yes	4
7*	9	F	W	No	No	Right head & eyes	Right tonic-clonic	4	Left > right occipital	PHT/3	No	11½
8	10	M	W	Yes	No	Right head & eyes	Right brachiofacial	12	Left occipital	CBZ/2½	No	7½
9	4	F	W	No	Yes	Left head & eyes	None	4	Right occipital & generalised S/W	CBZ/2	No	13½

*Seizure began with visual symptoms.
Pb: phenobarbitone; PHT: phenytoin; CBZ: carbamazepine; S/W: spike and wave.

Table 2   Frequency and clinical manifestations of subsequent seizures in four children with recurrence

Case No	Time after first seizure	Ictal manifestations	
		On waking	During sleep
1	4 Months		Brief tonic adversion of head and eyes, vomiting, loss of consciousness twice within 30 minutes
	5½ Months, 6 months	Brief tonic adversion of head and eyes, loss of consciousness, preceded and followed by headache	
2	6 Months (during medication)		Onset not witnessed. Loss of consciousness (30 minutes), on arousal, headache and vomiting
	14 Months	Brief generalised tonic, loss of consciousness	
	22 Months		Twitching of left mouth corner and clonic movement of left upper limb with loss of consciousness (45 minutes)
5	6 Months, 24 months (during reduced dose)	Brief tonic adversion of head and eyes, vomiting, loss of consciousness	
6	8 Months, 14 months	Brief complex partial seizure, post-ictal headache (10 minutes)	

within three years the EEG normalised and eventually the medication was reduced and finally discontinued.

Case 8   This 10 year old boy, generally in good health, complained of a sudden severe headache while in school. Some 20 minutes later he became confused and shortly thereafter lost consciousness. On examination in the paediatric ward two hours later he was still unconscious with deviation of the head and eyes to the right and fine clonic jerks of the right mouth angle and right upper extremity. An ictal EEG showed continuous bilateral slow spike and wave complexes, predominantly on the left. During EEG the spike activity disappeared completely after intravenous administration of 7 mg diazepam, with the appearance of drug-induced beta activity over the right hemisphere and post-ictal delta waves on the left, more prominent in the posterior region. The convulsions stopped, but the child remained unconscious. There were no signs of increased intracranial pressure. CT scan of the brain was normal. Twelve hours after onset of this episode the child regained consciousness, without any neurological deficit. Carbamazepine therapy, 600 mg per day, was instituted. EEG recordings made one day and then one week later still showed slow activity (theta and delta) over the left hemisphere, predominantly in the occipital region. In consecutive EEGs performed six weeks later and then twice a year there were rhythmic sharp and slow wave discharges in the left posterior region, inhibited by eye opening. Two and a half years later there was normalisation of the EEG, and treatment was discontinued gradually. He has remained free of seizures after withdrawal of medication for more than five years.

Discussion

The alarming feature in these nine children was the prolonged loss of consciousness with tonic deviation of either of eyes alone or of both head and eyes either preceded by headache or accompanied by vomiting in a previously healthy child. In all these cases an acute cerebral insult—for example, increased intracranial pressure, cerebrovascular accident, or focal encephalitis—was suspected, and the child was consequently subjected to emergency investigations, including lumbar puncture and CT of the brain, and was admitted as critically ill to a paediatric or neurosurgical intensive care unit. Case 1 was first diagnosed as having oedema of the left hemisphere, was initially treated with iv steroids and mannitol and received acyclovir several hours later when focal encephalitis was considered to be the probable diagnosis. In case 2 iv dexamethasone was given with the diagnosis of brain oedema. In two cases (5 and 6) toxic encephalopathy was also considered in the differential diagnosis. In no case, however, was the possibility of an idiopathic epilepsy even considered. In all these cases the rapid recovery occurring within less than 24 hours and then the finding of an occipital epileptic focus came as a surprise, ruling out the possibility of an acute cerebral insult. The finding of epileptic activity either during the episode (case 8) or within 24 hours thereafter (cases 3, 4, 6) revealed the true aetiology of the event. Hence the importance of an urgent EEG during the ictus in such cases. EEGs recorded shortly after the ictal events, however, may show focal occipital slowing and spikes. This, with the additional poverty of fast rhythms in the same focal areas, may encourage the diagnosis of underlying structural lesion. Subsequent development of occipital paroxysms clarifies the diagnosis. In case 1 the diagnosis of BOE was made only after four months when the child had a second seizure and the EEG performed at that time revealed the occipital spike-focus; indeed the course was benign. All the patients were eventually controlled by standard anticonvulsants. In five cases there was no recurrence whatsoever, and in the other four (cases 1, 2, 5, 6) there were subsequently a few seizures, none of them severe. In five cases (1, 3, 7, 8, 9) treatment has already been discontinued with no recurrence.

The seizures in BOE are often brief, especially when occurring on waking and sometimes even without loss of consciousness.[27] Prolonged seizures lasting from 45 minutes to three hours have been mentioned in some reports.[24 12] Panayiotopoulos[9] found in 16 patients that "consciousness was preserved throughout the ictus in four patients. In all the other children consciousness was impaired or lost either from the onset or during the course of the fits. Seizures lasted only a few minutes

(nine patients) or were prolonged for several hours (seven children)." A review of the eight cases described by Panayiotopoulos[12] reveals that the "duration of the seizure varies from seven minutes up to three hours, but usually lasts from 10 to 15 minutes." Among the seven cases reported in detail by Gastaut[2] the seventh case had, at the age of 4 years, a nocturnal seizure starting with adversion to the right, followed by nausea, vomiting, and diarrhoea before losing consciousness for over one hour. Newton and Aicardi[4] reported among their 16 cases one 8 year old girl who "had bouts of loss of consciousness lasting between a few minutes to two hours, most about 30 minutes."

Bickerstaff[13] has described impairment of consciousness in several patients with basilar artery migraine, but this was of slow onset and short duration and never sufficiently profound to render the patient unarousable. Furthermore, these patients had no seizures and no EEG abnormalities were mentioned. Prolonged confusional states associated with dizziness and disturbed sensorium with varying degrees of agitation, simulating toxic metabolic psychosis, encephalitis, or acute toxic encephalopathy were described by Gascon and Barlow[14] as a presenting feature in four patients with juvenile migraine, none of whom had epileptic seizures. EEG recordings obtained shortly after these episodes were either within normal limits or showed posterior or diffuse slow wave activity. None of the children reported as suffering from basilar artery migraine[15 16] showed protracted loss of consciousness but rather suffered from confusional states. Furthermore, these attacks were not associated with convulsive seizures nor with occipital spikewaves. With regard to the other accompanying symptoms, in basilar artery migraine the headache supervenes after the visual aura and the other symptoms of brain stem dysfunction whereas in our cases it heralded the seizures. Prodromal headache preceding epileptic seizures had been described by Penfield and Jasper.[17]

Ictal vomiting may be common in BOE[12 18]; interestingly in four of our nine cases vomiting occurred during the adversive phase of the seizure. Panayiotopoulos[9 12] has recently emphasised that adversion is common in BOE, particularly in nocturnal seizures. In all but one of our nine patients it was present and in 22

(35%) of the patients previously reported.[6] Indeed, contraversive seizures were the most prominent motor manifestation in this group of 62 patients. The clinician facing a child who has suddenly lapsed into a coma associated with focal features such as tonic deviation of either of eyes alone or of both head and eyes should be aware that besides an acute cerebral insult such symptoms may represent an occipital seizure disorder which may prove to be benign. The early performance of an EEG, preferably during the seizure, may clarify the issue. The finding of an occipital focus in such cases indicates a favourable outcome.

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